

## ***A Very Rare Case of a Retrospectively Diagnosed Iliocecal Neuroendocrine Tumor***

### **Retrospektif Olarak Tanı Konulmuş Oldukça Nadir Bir İleoçekal Nöroendokrin Tümör Vakası**

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#### **Abstract**

Neuroendocrine tumors usually cause symptoms – because of the release of vasoactive substances. Diagnosis is confirmed by detection of high levels of 5-hydroxyindoleacetic acid in urine. These typical symptoms may be absent, particularly when it originates from gastrointestinal tract. Gastrointestinal neuroendocrine tumors are silent; hence, they are difficult to diagnose in the initial stages but have good prognosis, if properly treated. We present the successful management of a patient who underwent right hemicolectomy and was postoperatively revealed to be a case of iliocecal neuroendocrine tumor on histopathologic examination of the specimen.

**Keywords:** Neuroendocrine tumor, iliocecal carcinoid tumor, hemicolectomy

#### **Öz**

Nöroendokrin tümörler vazoaktif maddelerin salınımından dolayı genellikle semptomlara yol açarlar. İdrarda yüksek düzeyde 5-hidroksi indol asetik asit belirlenmesiyle tanı doğrulanır. Özellikle gastrointestinal kanalda ortaya çıktığında bu tipik semptomlar olmayabilir. Gastrointestinal nöroendokrin tümörler sessizce gelişirler, bu nedenle başlangıç evrelerinde teşhis edilmeleri zordur. Ancak doğru bir şekilde tedavi edilirse iyi prognoz gösterirler. Bu çalışmada, sağ hemikolektomi uygulanan ve ameliyat sonrasında numunenin histopatolojik incelemesinde ileoçekal nöroendokrin tümörü olduğu ortaya çıkan bir hastanın başarılı yönetimi sunulmaktadır.

**Anahtar Kelimeler:** Nöroendokrin tümör, ileoçekal karsinoid tümör, hemikolektomi

#### **INTRODUCTION**

Neuroendocrine tumors, previously known as carcinoid tumors, are slow-growing tumors originating from neuroendocrine cells capable of metastasis (1, 2). Majority of them are found in the gastrointestinal tract (GIT); however, they can also be rarely seen in other organs (1, 3, 4). Neuroendocrine tumors in the ileum tend to be more aggressive and have poorer prognosis than those at other locations (5). They secrete approximately 40 bioactive substances, including serotonin, histamine, kallikreins, 5-hydroxytryptophan (6). When tumors are located in the GIT, their secretions reach the liver and are metabolized to nonactive substances and are excreted in bile and urine. However, secretions from neuroendocrine tumors located at sites other than GIT including metastases in the lungs, testis, and ovary directly reach the systemic circulation and cause carcinoid syndrome (1).

We present here the case of a patient with subacute intestinal obstruction, who underwent right hemicolectomy, that was postoperatively revealed to be a case of iliocecal neuroendocrine tumor on histopathologic examination of the specimen.

#### **CASE PRESENTATION**

The study protocol was approved by the institutional ethics committee, and informed consent was obtained from the patient.

A 70-year-old female weighing 66 kg, presented with complaints of intermittent pain in the abdomen and constipation since 2 years (suggestive of subacute intestinal obstruction), weakness, loss of appetite and weight loss since previous 6 months. She had type 2 diabetes and was on oral antidiabetics. She also had hypertension and hypothyroidism and was on appropriate medications. With these treatments, all parameters were well-controlled, except blood pressure which was 200/108 on admission. Her hemogram was within normal limits except for mild anemia (hemoglobin 11.6% g). She was negative for HbsAg and HIV. Her electrocardiogram showed mild left ventricular hypertrophy. Her barium follow through showed iliocecal stricture of the small intestine suggestive

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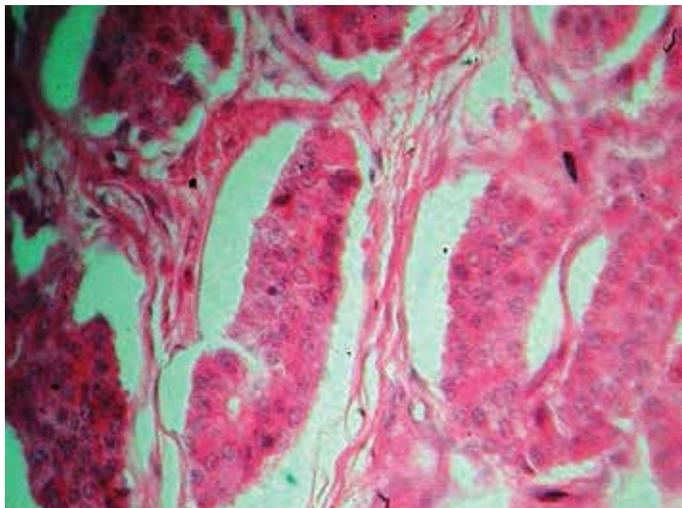


Figure 1. Histopathologic examination of specimen confirming the diagnosis of neuroendocrine tumor (H&E stain, 40×)

of possibility of iliocecal tuberculosis or malignancy. Considering the loss of appetite, weight loss, and radiological findings, she was started on anti-tuberculous treatment with no relief even after 2.5 months. Hence, she was prepped for planned right hemicolectomy.

To correct the mild anemia, she was given two units of packed cell volume, thus improving her hemoglobin to 13.6% g. To normalize blood pressure, she was administered 100 mg labetalol, a cardioselective beta blocker, twice a day in addition to a previous antihypertensive regime, which brought her blood pressure down to 146/88 mmHg in 6 days. She was preoperatively shifted to human insulin injection twice a day, which controlled her blood sugar level to normal limits. She received all antihypertensive and antithyroid drugs 4 h before surgery.

She was given combined spinal and epidural anesthesia with injection bupivacaine heavy 4 mL and epidural catheter for continuous epidural anaesthesia with sensory block up to T6. Surgery lasted for 3.5 h. A fall in blood pressure was noticed immediately after spinal anesthesia, requiring two doses of mephentermine injection 7.5 mg each; no such fluctuations in blood pressure were noted there after. After 1 h 25 min, the spinal anesthesia effect wore off, and epidural anesthesia was added with 20 mL 1% plain lignocaine + 10 mL 0.25% bupivacaine. Operative course was normal. After surgery, she was administered epidural analgesic with 150 µg buprenorphine injection + 100 mg tramadol injection diluted to 15 ml normal saline twice daily for 5 days, keeping epidural catheter in situ. She received insulin as per glucose level demand. Antihypertensive and eltroxin tablets were continued.

She was discharged on postoperative day 10 with continuation of all antihypertensive, antidiabetic, and tablet for hypothyroidism after ensuring adequate control of all ailments. She was instructed to follow up every week. Her iliocecal resected specimen was sent for histopathologic examination. Microscopic examination of the specimen showed solid, nesting, glandular masses of monotonous small circular cells with peripheral palisading; moderate amount of finely granular cytoplasm with small nucleoli and salt paper chromatin; and 2–3 mitotic activity per high power field, thus confirming the diagnosis of neuroendocrine tumor (Figure 1).

## DISCUSSION

Incidence of neuroendocrine tumors is 0.2–10 per 1,00,000 population (1,6). Incidental autopsy findings are common in asymptomatic patients (6). Although many organs are involved, majority of neuroendocrine tumors are found in the GIT (3-5), and approximately 70%–80% cases are those of GIT neuroendocrine tumors (1, 7). Of all secretions from neuroendocrine tumors, serotonin is a major bioactive substance (1, 2). Normally, 1% of dietary tryptophan is converted to serotonin, but in carcinoid, 70% of it is converted to serotonin, which in turn is converted to 5-hydroxyindoleacetic acid (5-HIAA) in the liver and secreted in urine. Elevated levels of 5-HIAA are markers of excess serotonin production, and 5-HIAA is an indicator for diagnosis of neuroendocrine tumors (1).

“Carcinoid crisis” results from sudden massive release of bioactive substances in circulation, causing wide fluctuations in blood pressure, hypotension, rarely hypertension, and tachycardia. GI hypermotility leading to diarrhea, wheezing, bronchospasm, and cyanosis are also common.

Our case presented with mechanical subacute intestinal obstruction due to tumor at iliocecal junction and did not have common symptoms suggestive of neuroendocrine tumors caused by the release of bioactive substances. Hence we could not have preoperative suspicion of neuroendocrine tumor. Our patient was retrospectively diagnosed with neuroendocrine tumor on histopathologic examination of the specimen, which explained the hypertension not responding to telmisartan and amlodipine during preoperative period that responded to cardioselective beta blocker labetalol. Careful preoperative examination and findings of fluctuating blood pressure mainly with hypertension not responding to routine antihypertensive drugs can raise the suspicion of a neuroendocrine tumor. Combined spinal and epidural techniques can safely be used after controlling all vital parameters, such as heart rate and blood pressure, with strict monitoring of blood pressure, intravenous fluids, and good postoperative analgesia by epidural route (8, 9).

Examination after 15 days showed blood pressure of 106/68 and after 7 days, blood pressure had decreased to 96/60. Hence, antihypertensive (Tablet Labetalol) agent was tapered off. This explains some partial effect of the release of vasoactive substances from tumors. Subsequently, her blood pressure was stable in normal limits.

## CONCLUSION

Gastrointestinal tract neuroendocrine tumors can present as subacute intestinal obstruction. Unless classical symptoms of neuroendocrine tumors are present, it is difficult to have preoperative suspicion, particularly in neuroendocrine tumors originating from the GIT. Omission of the antihypertensive tablet labetalol after some weeks postoperatively suggest that some effects of neuroendocrine secretions lead to an exaggeration of blood pressure effect before surgery. A well-controlled patient with neuroendocrine tumors and stable cardiovascular status can be safely managed with combined spinal and epidural techniques.

**Ethics Committee Approval:** Ethics committee approval was received for this study from the ethics committee of MIMSR Medical College (Date: 19.01.2015/MIMSR/EC/02/2015).

**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

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